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Idiopathic Non Cirrhotic Portal Hypertension with Portal Gastropathy: A Case Report

Barnali Bhuyan[1], Kunal Bhowmik[2], Ibandalin M. Shangpliang[3], Rintu Marak[4], Rikyntilin Wahlang[5]

[1]Unit Head, Department of Internal Medicine, Nazareth Hospital, Shillong, Meghalaya, India
Email: shyavak30[at]gmail.com

[2]Second Year DNB, Department of Internal Medicine, Nazareth Hospital, Shillong, Meghalaya, India
Email: bicky_kunal[at]yahoo.co.in

[3]Consultant, Department of Internal Medicine, Nazareth Hospital, Shillong, Meghalaya, India
Email: jboadita[at]gmail.com

[4]Consultant, Department of Internal Medicine, Nazareth Hospital, Shillong, Meghalaya, India
Email: r_marak84[at]yahoo.com

[5]Senior Medical Officer, Department of Internal Medicine, Nazareth Hospital, Shillong, Meghalaya, India
Email: rikyntiwahlang[at]gmail.com

Running Title: *Idiopathic portal hypertension*

Abstract: ***Idiopathic portal hypertension (IPH) is a rare disorder characterized by portal hypertension without a recognizable cause such as cirrhosis. It is clinically characterized by splenomegaly, features of hypersplenism, and increased portal or splenic venous pressure. Despite reported cases of IPH, the entity still remains fairly under diagnosed. We are reporting a case of a 27 years old female, with no significant comorbidities and addiction history, who presented with complains of vomiting coffee brown substance and passing black colored stool. Her abdominal CECT and ultrasonography findings were consistent with sequelae of portal hypertension, with normal hepatic architecture and splenomegaly. Transient elastography confirmed the absence of cirrhosis. Esophagogastroduodenoscopy revealed esophageal varices and band ligation was done. She symptomatically improved and was later discharged on nonselective beta blocker therapy. This case highlights the unique possibility of finding features of hypersplenism without any underlying hepatic pathology.***

Keywords: idiopathic portal hypertension, variceal hemorrhage, non-cirrhotic portal hypertension, case report

1. Introduction

Idiopathic portal hypertension (IPH) is an important disease entity characterized by the presence of portal hypertension due to intrahepatic or prehepatic lesions in the absence of cirrhosis. It is a common cause of noncirrhotic portal hypertension and has been widely reported worldwide. Other terms for this condition used include hepatoportal sclerosis, obliterative venopathy and sometimes Banti's syndrome when accompanied by splenomegaly and features of hypersplenism. (1)

Despite many documented cases of IPH, India lacks a national registry and nationwide studies to determine the incidence of the disease. Studies conducted in and prior to 1980s found an incidence of 7.9–46.7%, peaking in the third and fourth decades of life with a male predominance as opposed to female predominance in other parts of the world. (2) In 2006, Pande et al. conducted a clinical profile of 366 patients in a tertiary care center in New Delhi and found 98% of patients with IPH were from the low-socioeconomic class. (3)

A number of causative hypothesis including infections, prothrombotic state, xenobiotic exposure, immunological abnormalities have been proposed till date but no definitive consensus has been reached.

IPHpatients usually present with episodes of gastrointestinal hemorrhage, palpable mass in the left hypochondrium, anemia and consequences of hypersplenism. Laboratory tests usually reveal a near normal liver function sometimes along with pancytopenia. Imaging studies establish features of portal hypertension whereas hepatic echotexture is usually normal. Development of ascites, jaundice, and hepatic encephalopathy is uncommon and may be seen only after an episode of gastrointestinal hemorrhage. Esophagogastric varices have been reported in 85%-95% of patients with IPH and approximately 70% present with a major variceal hemorrhage. (2)

Medical management usually comprises of blood transfusions, intravenous fluids, prophylactic antibiotics and use of vasoactive drugs such as somatostatin, terlipressin or octreotide. Endoscopic band ligation in combination with medical therapy has proven effective in controlling acute variceal bleed and preventing rebleeding. Non selective beta blockers are advocated as a preventive measure in portal hypertension secondary to cirrhosis but no effective consensus has been reached in IPH patients. (2) Transjugular intrahepatic portosystemic shunt can be considered for uncontrollable hemorrhage or recurrent episodes despite endoscopic intervention. (4) However there is a lack of management guidelines in the specific scenario of portal hypertension without cirrhosis.

2. Case Report

A 27 years old female presented to the emergency department with complains of vomiting copious amounts of coffee brown colored substance on 2 occasions the day before and passing black colored stool since then.

She had a similar episode in 2017 requiring hospitalization but no documents were available.

She had no other complaints and no known significant comorbidities.

She had no history of any addiction and was not on any long-term medications.

On examination, she had pallor. Abdominal examination revealed non tender splenomegaly approximately 7 cm below the left subcostal margin. Rest of her examination was normal.

Her initial blood reports showed 6.1 g/dL hemoglobin, 4320 WBCs/cumm and 208000 platelets/cumm. Her liver function test showed atotal bilirubin of 1.7 g/dL with 1.5 g/dL being the unconjugated fraction, mildly elevated transaminases and a normal albumin globulin ratio. Her coagulation profile and other reports were grossly normal. She was initially kept nil per oral and received tranexamic acid, pantoprazole and blood transfusion. Abdominal ultrasonography showed a normal hepatic echotexture with increased echogenicity of intrahepatic portal vein branches and a moderately enlarged spleen of 17.8 cm (Figure 1). CECT abdomen showed splenomegaly with 874 splenic index along with perisplenic, perigastric, peripancreatic, umbilical and periportal varices (Figure 2). Transient elastography showed a median hepatic stiffness of 3.7 kPa. Esophagogastroduodenoscopy showed three columns of grade III varices in the distal third of esophagus with no active bleeding along with portal gastropathy. She was started on propranolol prophylactically and endoscopic band ligation was done thereafter (Figure 3). Following that she symptomatically improved and was eventually discharged after 6 days of hospitalization.

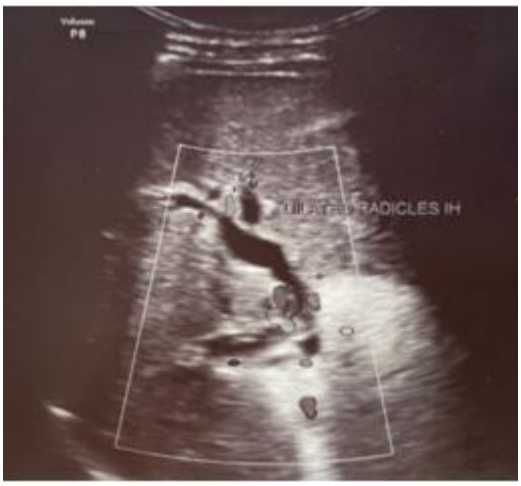

Figure 1: Ultrasound showing normal hepatic echotexture

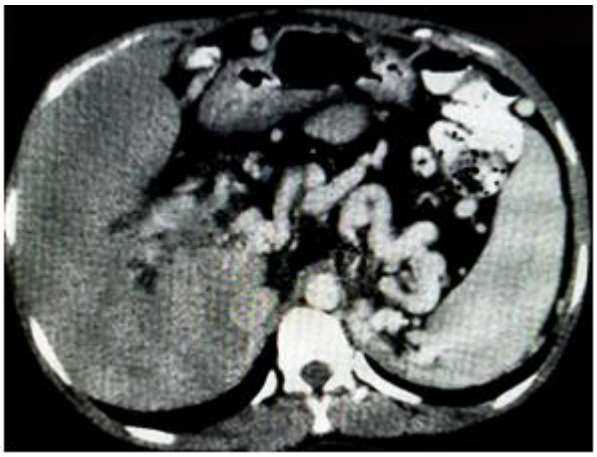

Figure 2: CT portalphase showing dilated splenic vein with dilations suggestive of varices

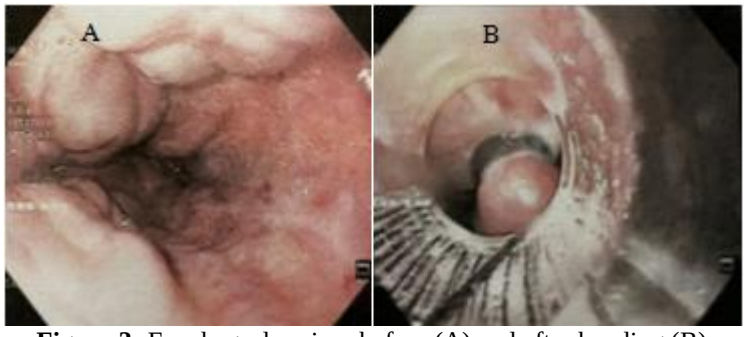

Figure 3: Esophageal varices before (A) and after banding (B)

3. Discussion

Hematemesis and malena are frequently encountered presentations in the medical emergency, most of which are attributed to ulcers or variceal hemorrhage. Even though varices secondary to portal hypertension is commonly encountered in patients with decompensated liver disease, there have been increasing reports of idiopathic non cirrhotic portal hypertension with features of hypersplenism.

In IPH, portal venous blood flow into the liver and the diameter of portal vein trunks are increased in contrast to an almost normal portal venous blood flow in patients with portal hypertension secondary to cirrhosis, as clearly evidenced in this case. Multiple hypotheses regarding its etiopathogenesis have been made till date, with more recently increased serum levels of vascular adhesion molecule-1 (VCAM-1) and their expression on the

endothelium of hepatic vessels in IPH patients being implicated in the development of portal venous sclerosis ultimately resulting in presinusoidal portal hypertension. (5) But a definitive etiology is yet to be established. IPH patients mostly present with hematemesis which can quickly contribute to hemodynamic instability of the patient if not addressed aggressively. Till date, combined medical and endoscopic intervention has been associated with the best outcomes in such patients and the management follows the protocols of variceal bleed secondary to cirrhosis. However, more insight into IPH is required establishing the etiopathogenesis and identifying risk factors which might lead to emergence of new therapeutic guidelines as well as preventive measures and provide clear evidence regarding the benefits of interventions like prophylactic antibiotics in the setting of variceal bleed without concomitant cirrhosis. We are reporting this case hoping to drive the medical community to look beyond the common causes of upper gastrointestinal bleed and encourage further research into the field.

4. Conclusion

IPH remains a common disease entity with a possible multifactorial etiology, prevalent in certain geographical areas and populations. The clinical presentation is usually with splenomegaly and/or complications of portal hypertension and hypersplenism. Diagnosis is based on laboratory and radiological findings. Patients with IPH have a fairly good prognosis and normal life expectancy if managed appropriately. Hence, more research is required in this field to establish management guidelines specific to this condition and possibly adopt preventive measures.

Conflicts of Interest and Source of Funding: None declared

References

[1] Bodh V, Chawla Y. Noncirrhotic intrahepatic portal hypertension. Clin Liver Dis.2014; 3 (6): 129–32.
[2] Sarin SK, Kumar A, Chawla YK, Baijal SS, Dhiman RK, Jafri W, et al. Noncirrhotic portal fibrosis/idiopathic portal hypertension: APASL recommendations for diagnosis and treatment. Hepatol Int.2007 Oct 15; 1 (3): 398–413.
[3] Pande C, Kumar A, Sarin S. Non-Cirrhotic Portal Fibrosis: A Clinical Profile of 366 Patients. Am J Gastroenterol.2006; 101 (p S191).
[4] Sarin SK, Kapoor D. Non-cirrhotic portal fibrosis: Current concepts and management. J Gastroenterol Hepatol.2002; 17 (5): 526–34.
[5] Nakanuma Y, Kouda W, Nakano T, Uneno K, Tachibana S, Araki I. A case report of early idiopathic portal hypertension. Pathol Res Pract.2001; 197 (11): 759–63.